Síndrome de vasculitis: diagnóstico y tratamiento

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Síndrome de vasculitis: diagnóstico y tratamiento
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Resumen
En pacientes con enfermedad del tejido conjuntivo, la lesión vascular inducida por síndromes de vasculitis primaria o secundaria puede provocar disfunción orgánica debido a la pérdida del suministro de nutrientes de la sangre. Dichos síndromes de vasculitis pueden ser refractarios al tratamiento y fatales. La nomenclatura y la definición de los síndromes de vasculitis se han revisado recientemente y las guías de práctica clínica para enfermedades asociadas con el síndrome de vasculitis están evolucionando. La presente revisión proporciona una visión general de los síndromes de vasculitis desde el punto de vista del diagnóstico y el tratamiento.

Palabras clave: diagnóstico, nomenclatura, terapia, síndrome de vasculitis
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1. Introducción
La vasculitis sistémica se divide en dos categorías principales: síndrome de vasculitis primaria y síndrome de vasculitis secundaria. La primera es un tipo de vasculitis causada por la inflamación de los vasos sanguíneos, mientras que la segunda es inducida por las condiciones subyacentes, que incluyen enfermedades del tejido conectivo, tumores, infecciones y alergias a medicamentos. El síndrome de vasculitis primaria y el síndrome de vasculitis secundaria asociado a enfermedad del tejido conectivo se incluyen dentro del concepto de enfermedad autoinmune sistémica. La etiología de todos los trastornos de vasculitis es esquiva. Los síndromes de vasculitis primaria se clasifican según el tamaño patológicamente confirmado de los vasos afectados. Los planes de tratamiento están diseñados para adaptarse a las respectivas enfermedades. La Conferencia de Consenso de

Chapel Hill (CHCC) en 1994 formó la base para la categorización de las enfermedades antes mencionadas.1 Sin embargo, casi 20 años después, el CHCC se reunió nuevamente para definir los síndromes de vasculitis primaria y secundaria que no se definieron de otra manera en su reunión de 1994. Otro propósito de la conferencia fue el reexamen y unificación de la nomenclatura de nuevos tipos de síndromes de vasculitis y sus enfermedades asociadas (Tabla 1). 2 Esta revisión proporciona una visión general de esta nueva nomenclatura.

tabla 1
Definición de vasculitis adoptada por la Conferencia de Consenso de Chapel Hill de 2012 sobre la nomenclatura de vasculitis

Vasculitis de grandes vasos (LVV)
Arteritis de Takayasu (TAK) Vasculitis que afecta a las grandes arterias con más frecuencia que otras vasculitis. Las grandes arterias son la aorta y sus principales ramas. Arteria de cualquier tamaño puede verse afectada
Arteritis de células gigantes (ACG)
Vasculitis de vasos medianos (MVV)
Poliarteritis nodosa (PAN) Vasculitis que afecta predominantemente a las arterias medias definidas como las principales arterias viscerales y sus ramas. Cualquier arteria de tamaño puede verse afectada. Los aneurismas inflamatorios y las estenosis son comunes
Enfermedad de Kawasaki (EK)
Vasculitis de pequeños vasos (SVV) Vasculitis que afecta predominantemente a los vasos pequeños, definidos como pequeñas arterias intraparenquimatosas, arteriolas, capilares y vénulas. Las arterias y venas medianas pueden verse afectadas.
Vasculitis asociada a ANCA (VAA)
Poliangitis microscópica (MPA) Vasculitis necrosante, con pocos o ningún depósito inmunitario, que afecta predominantemente a vasos pequeños (es decir, capilares, vénulas, arteriolas y arterias pequeñas), asociada con mieloperoxidasa (MPO) ANCA o proteinasa 3 (PR3) ANCA. No todos los pacientes tienen ANCA. Agregue un prefijo que indique reactividad ANCA, por ejemplo, MPO-ANCA, PR3-ANCA, ANCA negativo
Glanulomatosis con poliangitis (GPA) a (antigua glanulomatosis de Wegener)
Granulomatosa eosinofílica con poliangeítis (EGPA) a (antiguo síndrome de Churg-Strauss)
Vasculitis por inmunocomplejos
Enfermedad de la membrana basal antiglomerular (Anti-GBM) a (antiguo síndrome de Goodpasture) Vasculitis con depósitos moderados a marcados en la pared de los vasos de inmunoglobulina y/o componentes del complemento que afectan predominantemente a los vasos pequeños (es decir, capilares, vénulas, arteriolas y arterias pequeñas). La glomerulonefritis es frecuente.
Vasculitis crioglobulinémica (CV)
Vasculitis IgA (IgAV) a (antigua púrpura de Henoch-Schönlein)
Vasculitis urticarial hipocomplementémica (HUV) (vasculitis anti-C1q)
Vasculitis de vaso variable (VVV)
Enfermedad de Behçet (EB) Vasculitis with no predominant type of vessel involved that can affect vessels of any size (small, medium, and large) and type (arteries, veins, and capillaries)
Cogan's syndrome (CS)
Single organ vasculitis (SOV)

Cutaneous leukocytoclastic angiitis Vasculitis in arteries or veins of any size in a single organ that has no features that indicate that it is a limited expression of a systemic vasculitis. The involved organ and vessel type should be included in the name (eg, cutaneous small vessel vasculitis, testicular arteritis, central nervous system vasculitis). Vasculitis distribution may be unifocal or multifocal (diffuse) within an organ. Some patients originally diagnosed as having SOV will develop additional disease manifestations that warrant redefining the case as one of the systemic vasculitides (eg, cutaneous arteritis later becoming systemic polyarteritis nodosa, etc.)
Cutaneous arteritis
Primary CNS vasculitis
Isolated aortitis
Others
Vasculitis associated with systemic disease
Lupus vasculitis Vasculitis that is associated with and may be secondary to (caused by) a systemic disease. The name (diagnosis) should have a prefix term specifying the systemic disease (eg, rheumatoid vasculitis, lupus vasculitis, etc.)
Rheumatoid vasculitis
Sarcoid vasculitis
Relapsing polychondritis vasculitis
Others
Vasculitis associated with probable etiology
Hepatitis C virus-associated cryoglobulinemic vasculitis Vasculitis that is associated with a probable specific etiology. The name (diagnosis) should have a prefix term specifying the association (eg, hydralazine-associated microscopic polyangiitis, hepatitis B virus-associated vasculitis, hepatitis C virus-associated cryoglobulinemic vasculitis, etc.)
Vasculitis asociada al virus de la hepatitis B
Aortitis asociada a sífilis
Vasculitis por inmunocomplejos asociada a fármacos
Vasculitis asociada a ANCA asociada a fármacos
Vasculitis asociada al cáncer
Otros
Abrir en una ventana separada
a La enfermedad a la que se cambió una nomenclatura (Las nuevas nomenclaturas se expresan en negrita). Revisado de la referencia 2 .
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2. Concepto de Síndrome de Vasculitis para el Diagnóstico
La base fisiopatológica de las características clínicas de la vasculitis primaria está relacionada con el hecho de que los vasos sanguíneos suministran oxígeno para mantener la función de los órganos sólidos y el intersticio. Como resultado de la inflamación de los vasos sanguíneos, el sangrado causado por la ruptura de las estructuras vasculares (secundario a la inflamación) o la interrupción del flujo sanguíneo puede provocar una disfunción orgánica. Esto ocurre en el área local de la lesión vasculítica así como en los órganos viscerales y el intersticio en el área de perfusión periférica. Con respecto al diagnóstico del síndrome de vasculitis primaria, es importante sistematizar los síntomas localizados individuales que, a primera vista, parecen ser independientes. El médico debe evaluar si tales síntomas localizados individuales son o no el resultado de la isquemia causada por la inflamación de los vasos sanguíneos o debido a síntomas que surgen como resultado de una hemorragia. Por lo tanto, en lugar de tener en mente un diagnóstico de

vasculitis individual subdividida, es más importante que los médicos comprendan el proceso de diagnóstico del síndrome de vasculitis (Figura 1) y los síntomas clínicos de vasculitis.

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Figura 1
Aproximación al diagnóstico del síndrome de vasculitis. TAK , arteritis de Takayasu; GCA , arteritis de células gigantes; PAN , poliarteritis nodosa; MPA , poliangitis microscópica; EGPA , granulomatosis eosinofílica con poliangitis; GPA , granulomatosis con poliangitis; aGBM , enfermedad de la membrana basal antiglomerular; Ig AV , vasculitis IgA; CV , vasculitis crioglobulinémica; VD , vasculitis reumatoide. Revisado de la referencia 3.
Kawasaki disease is excluded in this diagnostic procedure because the diagnostic procedure for Kawasaki disease is based solely on characteristic clinical signs and symptoms

Go to:
3. Diagnosis of Vasculitis Syndromes
Clinical features of primary vasculitis syndromes can be classified largely into two categories: systemic signs/symptoms caused by inflammation and localized visceral signs/symptoms specific to the affected organs (Table 2).3 Signs/symptoms that could evoke the possibility of vasculitis syndrome are described as follows.

Table 2
Visceral manifestations of large-/medium-vessel arteritis and small-vessel vasculitis

I. Visceral manifestation of large-/medium-sized vessel	
Common carotid artery	Dizziness, headache, syncope
Maxillary artery	Jaw claudication
Ophthalmic artery	Visual loss, blindness
Subclavian artery	Numbness & cold sensation of upper extremities, easy fatigability, difference in blood pressure between right and left arm, pulse deficit
Renal artery	Hypertension, renal failure
Mesenteric artery	Ischemic colitis (abdominal pain melena etc.)
Coronary artery	Angina pectoris, myocardial infarction
Pulmonary artery	Cough, bloody sputum, dyspnea
II. Visceral manifestation of small-sized vessel arteritis	
Skin	Livedo reticularis, subcutaneous nodules, purpura, skin ulcer, finger/toe-tip necrosis
Peripheral nerve	Mononeuritis multiplex
Muscles	Myalgia
Joints	Arthralgia
Kidney	Necrotizing crescentic glomerulonephritis
Gastrointestinal tract	Gastrointestinal ulcer, gastrointestinal hemorrhage
Heart	Myocarditis, arrhythmia
Lung	Interstitial pneumonitis, pulmonary alveolar hemorrhage
Serous membrane	Pericarditis, pleuritis
Eye	Retinal hemorrhage (visual loss), scleritis

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Revised from Ozaki et al. 3.

3.1. Systemic manifestations
Fever of unknown origin (FUO): In many cases, the patient experiences a high, spiking fever of 38-39°C.
Weight loss caused by inflammation.
Weakness, general malaise.
Arthralgia, muscle pains: This systemic manifestation indicates that vasculitis has spread systemically, while localized manifestations result from insufficient supply of local blood flow caused by the inflammation of blood vessels. Histological study with biopsy is recommended in cases of localized myalgia.
The aforementioned systemic symptoms are frequently seen among elderly people with malignant tumors. Therefore, during the process of diagnosis for vasculitis syndrome, it is recommended to also conduct screening examinations for malignant tumors.4

3.2. Local symptoms
Local symptoms are featured by the simultaneous (or sequential) appearance of symptoms from different affected organs in a patient with vasculitis syndrome.

Visceral Signs/Symptoms of Large- and Medium-Vessel Vasculitis. As large to medium-sized blood vessels run from the aorta to the organs, the signs/symptoms of vasculitis result from injury to organs supplied by the affected vessels. These signs/symptoms include pulse deficit, jaw claudication, loss of vision, and acute abdomen. Especially in large-vessel vasculitis, clinicians should be aware of the possibility that any size of artery may be affected, because a decrease in aortic blood pressure can result in compromise of blood flow to all downstream arteries.
Visceral signs/symptoms of Small-Vessel Vasculitis. In cases with skin rash, the so-called palpable purpura is a distinguishing feature that frequently develops in the lower limbs. Mononeuritis multiplex is a clinical manifestation of vasculitis of medium to small arteries that feed the affected nerves. In the early stage, it may manifest as sensory disturbance, such as hyperesthesia or hypoesthesia, and, when progressive, leads to motor disturbance that may result in drop hand or foot. The clinical features of small-vessel vasculitis in the kidney include those of nephritis, such as hematuria, proteinuria, and cylindruria. In some cases, pulmonary alveolar hemorrhage caused by arteriolitis or venulitis in the lungs develops with bloody and foamy sputum.
3.3. Laboratory findings and imaging studies
General Laboratory Findings: Acute inflammatory markers, such as erythrocyte sedimentation rate (ESR), CRP, and white blood cell (WBC) count, may be present. In cases with eosinophilic granulomatosis with polyangiitis (EGPA), a significant increase in eosinophil is observed. At times, high level of plasma renin activity or HBV antigen positivity can be seen in a patient with polyarteritis nodosa (PAN).
Urinalysis: Even in its early stage, proteinuria, erythruria, leukocyturia, and cylindruria can be found in patients with microscopic polyangiitis (MPA). However, in PAN and granulomatosis with polyangiitis (GPA), there are cases in which abnormal urinary findings appear over time rather than immediately.
Biochemical markers: In MPA, PAN, and GPA, renal dysfunction, such as elevation of serum creatinine, a rise in BUN, and a decline in creatinine clearance, can be observed. As interstitial pneumonia is one of representative clinical features in MPA, a rise in KL-6 can also be seen.

Antineutrophil cytoplasmic antibody (ANCA): In MPA and EGPA, it is common (50%-80% of cases) to detect myeloperoxidase (MPO)-ANCA (perinuclear [p] staining pattern with an indirect immunofluorescent [IIF] assay, p-ANCA). Antineutrophil cytoplasmic antibody testing is usually negative in PAN (<20% ANCA positive). In GPA, proteinase 3 (PR3)-ANCA (cytoplasmic (c) staining pattern with IIF assay, c-ANCA) is positive (90%). In recent years, genetic factors,5 biological environment factors (the induction of neutrophil extracellular traps [NETs] from neutrophil caused by bacterial infections),6 scientific environment factors (antithyroid drugs, such as propylthiouracil,7 atmospheric environmental chemicals such as silica) have been presumed as causes of ANCA positivity. Detailed history taking via an interview is highly important to distinguish primary vasculitis from genetic or environmental factor-associated vasculitis in a patient with positive ANCA and equivocal clinical features of vasculitis.

Antiglomerular basement membrane (GBM) antibody: Especially in cases exhibiting RPGN and/or alveolar hemorrhage, it is essential to suspect anti-GBM disease. Serum anti-GBM antibody is specific to laboratory findings for anti-GBM disease. Although there are various methods to detect anti-GBM antibody, such as the IIF assay, hemagglutination assay, radioimmunoassay (RIA) method, and enzyme-linked immunoassay (ELISA) method, the RIA and ELISA method have superior sensitivity (>95%) and specificity (>97%).8

Imaging study: Plain radiographs of the chest and paranasal sinuses, ultrasonography, CT, MRI, and thermography have been used to confirm abnormal vascular wall structure and blood flow dysfunction. If acute-phase Takayasu disease (TAK) is present, aortic wall thickening will be densely stained with gadolinium on contrast-enhanced MRI. As MRA can clearly outline whether or not there are any irregularities, contractions, or occlusion of the vascular walls, this imaging modality is often useful for interpretation of symptoms related to ischemia. Further, it may be possible to localize aortic inflammation using 18F-fluorodeoxyglucose-positron emission tomography (FDG-PET). Increased glycolytic metabolism in inflammatory tissues or malignant tumors is the rationale behind the common use of FDG-PET.9 Diagnostic imaging studies of the thorax (X-rays, CT, MRI) may show interstitial infiltrative shadows in the lung field in a patient with MPA. Granulomatous lesions can also be accompanied by infiltrative shadows in patients with EGPA and GPA. PAN can cause multiple microaneurysms and/or contractions in association with inflammation of medium-sized and small arteries. Aneurysms can be frequently detected in the branches of the abdominal aorta (eg, renal, mesenteric, and hepatic arteries) and can be confirmed via angiography. However, aneurysms are usually not detected during the acute phase. In some cases, it is possible to detect impaired blood flow using MRA or the ultrasonic Doppler method. However, in Japan, some physicians tend to avoid conventional angiography because of its invasiveness and because PAN can typically be diagnosed via biopsy.

Physiological tests: electrocardiography (ECG), pulse-wave detection, electroencephalography (EEG), electromyography (EMG), and nerve conduction studies are important for the diagnosis of vasculitis syndrome and to classify its severity. EMG and nerve conduction studies are crucial for the diagnosis of latent vasculitis syndrome and to identify appropriate biopsy sites.

3.4. Tissue biopsy

With regard to small-vessel vasculitis, it is essential to test for ANCA and immune complexes. However, these are definitive factors for diagnosis; rather, tissue biopsy is the most important diagnostic method for vasculitic syndrome. If it is necessary to differentiate ANCA-associated vasculitis (AAV) from immune complex-mediated vasculitis, it is preferable to prepare the materials for IIF staining in advance before conducting the tissue

biopsy. Because the primary lesions of TAK are in the aorta and those of Kawasaki disease (KD) are in the coronary arteries, it is not possible to perform a biopsy for diagnosis of these diseases. Hence, in such cases, diagnostic imaging study plays a central role in diagnosis. For other primary vasculitis syndromes, biopsy of the affected vessels is the most useful method for diagnosis.

Some considerations with regard to tissue biopsy for systemic vasculitis are as follows:

Giant cell arteritis (GCA): A biopsy of the temporal artery is essential for diagnosis. As noncontiguous segmental lesions develop in GCA, it is crucial to prepare serial sections of biopsy specimens.10
PAN: Although the typical histological finding of PAN is necrotizing angiitis, which is characterized by fibrinoid necrosis in the tunica media, old and new lesions are often observed within the same tissues. By definition, PAN does not include inflammation of the arterioles, venules, and capillaries, meaning that PAN is not associated with glomerulonephritis.
MPA: The typical histological finding of MPA is necrotizing angiitis in arterioles and capillaries in the kidneys, indicating necrotizing crescentic glomerulonephritis.
GPA: A finding of necrotizing granulomatous lesions with giant cells in lesion sites of the upper respiratory tract (eg, nose, paranasal sinuses, and soft palate) is useful for early diagnosis. Various patterns of glomerulonephritis can also be found in kidney biopsy specimens.
EGPA: Biopsy specimens from peripheral nerves, muscles, and lungs with infiltration show vasculitides and granulomas with prominent invasion with eosinophils.
Anti-GBM Disease: Diffuse crescentic glomerulonephritis is a distinctive feature of this disease. IIF staining can show deposition of IgG and C3 along the glomerular capillary walls.
IgA vasculitis (IgAV): Necrotizing angiitis can be observed with blood vessels in the area from the papillary layer to the reticular layer of the skin. Using immunofluorescent staining, in accordance with the areas where necrotizing angiitis has occurred, deposition of IgA in areas from the vascular endothelial cells to the vascular lumen can be observed.
As mentioned above, to diagnose a typical vasculitis syndrome, clinicians must collect patient information via interview, physical examinations, laboratory testing, imaging studies, and histopathological examination of biopsy specimens. This allows establishment of a diagnosis of vasculitis syndrome based on the classification criteria. After diagnosis, treatment should commence as soon as possible. However, it may require considerable time to establish a definitive diagnosis of vasculitis syndrome. During the diagnosis process, some patients may experience worsening in their condition. Therefore, in cases of vasculitis syndrome, rapid testing and treatment is necessary, hospitalization should be considered to increase the speed of this process. Further, referral or transfer to specialized centers is highly recommended if vasculitis syndrome is suspected.

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4. Treatment of Vasculitis

To prevent irreversible organ failure caused by the progression of vasculitis, it is important to use immunosuppressant drugs to reduce inflammation as soon as possible. It is also critical to maintain blood flow through the affected vessels.

In most cases, the first-line drug for the treatment of vasculitis syndrome is a corticosteroid. Some of the treatment methods for typical vasculitis syndrome are listed below:

4.1. Treatment of large-vessel vasculitis
Corticosteroid is effective for most patients with TAK and GCA. Strong anti-inflammatory effect has enabled a physician to begin administration with steroids in the early stages of the disease. However, some cases of vasculitis are resistant to corticosteroids. Therefore, attempts have been made to develop new methods of treatment using immunosuppressants to control such cases of steroid-resistant vasculitis (Figure 2).11

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Figure 2
Protocol for medical treatment of Takayasu arteritis. PSL, prednisolone; MTX, methotrexate; PO, oral administration; IV, intravenous injection; CsA, cyclosporin A; AZP, azathioprine; MMF, mycophenolate mofetil. Revised from reference 3

4.2. Treatment for medium-vessel vasculitis
Polyarteritis nodosa is the representative medium-vessel vasculitis. Based on the classification of vasculitis syndrome by the CHCC in 1994, MPA was separated from PAN. Since then, the number of reported cases of PAN has decreased. The response to corticosteroid therapy is better with PAN than with MPA. However, in patients with organ failure, combination therapy with immunosuppressive agents is desirable (Figure 3).

Un archivo externo que contiene una imagen, ilustración, etc. El nombre del objeto es JGF2-18-72-g003.jpg
Figure 3
Induction therapy for remission in polyarteritis nodosa. Revised from reference 3. mPSL, methylprednisolone; PSL, prednisolone; IVCY, intravenous cyclophosphamide; CY, cyclophosphamide

4.3. Treatment for small-vessel vasculitis
Microscopic polyangiitis is a representative of small-vessel vasculitis with necrotizing angiitis of small vessels (capillaries and arterioles). In many cases, lungs and kidneys are the major target organs. The incidence of ANCA positivity is high (~80%) in patients with MPA. In Japan, cases of MPO-ANCA are more predominant than those of PR3-ANCA.12 In Western countries, there are more positive cases of proteinase 3 (PR3)-ANCA in AAV. Treatment reports regarding AAV in Western countries mainly involve cases with positive PR3-ANCA.13 Three groups of the Ministry of Health and Labor's Research on Measures for Intractable Diseases Project in Japan jointly established the clinical practice guidelines for ANCA-associated vasculitis with MPO-ANCA-positive vasculitis. Especially in the JMAAV protocol, AAV is classified into three separate categories: mild cases (renal limited type, pulmonary fibrosis type), severe cases (systemic vasculitis, interstitial pneumonitis with glomerulonephritis, and RPGN type), and most severe cases (diffuse alveolar hemorrhage, intestinal perforation type). They used three different protocols for remission induction therapy using immunosuppressive agents according to the degree of severity. The main drugs used in the combination therapy are corticosteroids and cyclophosphamide.14 Furthermore, in Japan, other than the aforementioned standardized drugs for remission

induction therapy, various treatments (eg, intravenous immunoglobulin therapy for cases with peripheral nerve involvement caused by steroid-resistant EGPA15; or treatment with the anti-CD20 antibody, rituximab,16 in GPA or MPA that is resistant to the standard therapy with cyclophosphamide and corticosteroids) are permitted under the medical care insurance system.

Vasculitis syndrome is a rare disease of unknown etiology. In addition to advances in knowledge regarding the pathogenesis of vasculitis, we believe that large multicenter clinical trials in Japan could help define the etiology and optimal treatment for vasculitis syndrome.

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Conflict of Interest

The authors have stated explicitly that there are no conflicts of interest in connection with this article.

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Notes

Okazaki T, Shinagawa S, Mikage H. Vasculitis syndrome—diagnosis and therapy. J Gen Fam Med. 2017;18:72–78. https://doi.org/10.1002/jgf2.4 [PMC free article] [PubMed] [Google Scholar]

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